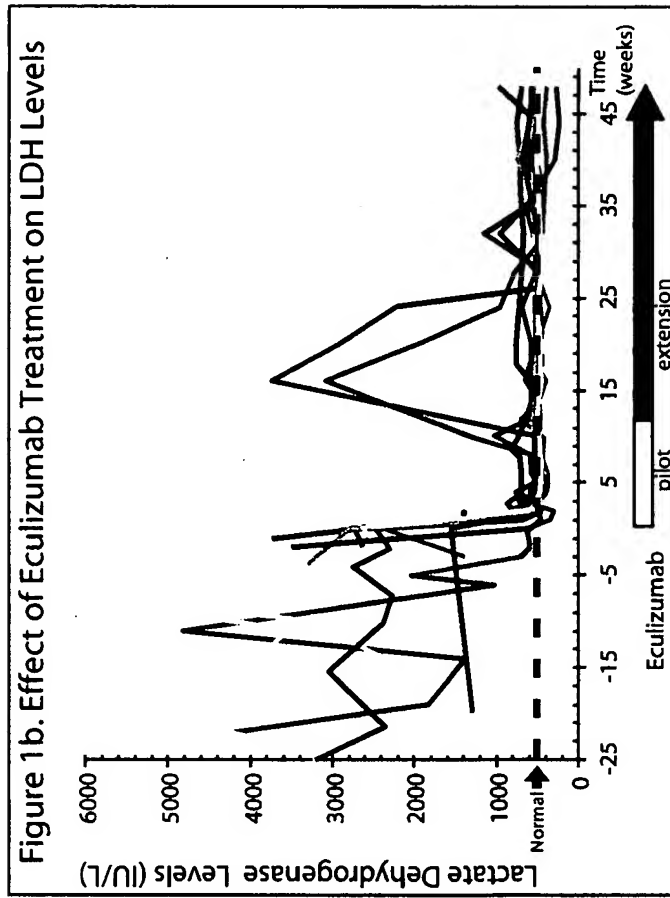


**Figure 1A. Biochemical Parameters of Hemolysis
During Eculizumab Treatment (mean values)**

	Normal range	Pre- study	Baseline	Week 12	1 year	P- value
LDH (IU/l)	150 - 480	3111	3017	564	547	0.002
AST (IU/l)	10 - 40	N.A.	76	28	31	0.028
Bilirubin (umol/l)	3 - 15	26	28	30	30	N.S.
Haptoglobin (g/dl)	0.5 - 2.0	<0.06	<0.06	0.07	0.17	N.S.
Hemoglobin (g/dl)	13.5 - 18 11.5 - 16	10.0	10.5	10.1	10.7	N.S.
Reticulocytes (x10 ⁹ /l)	20 - 80	161	157	222	197	N.S.



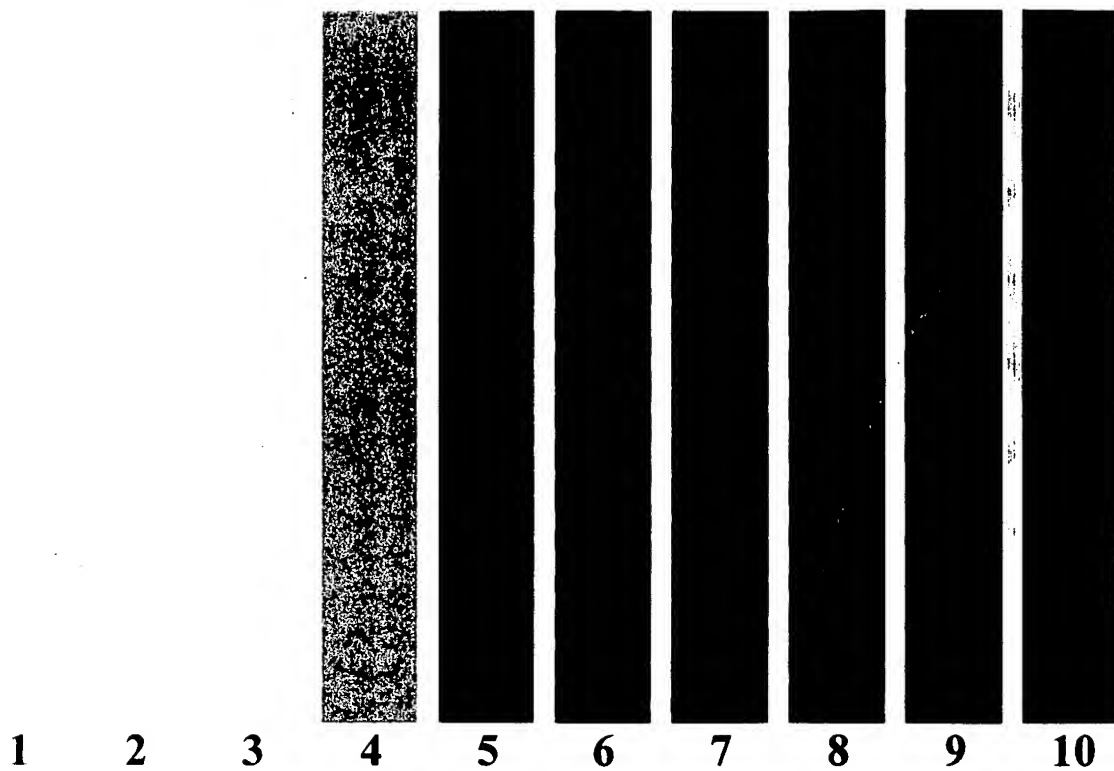
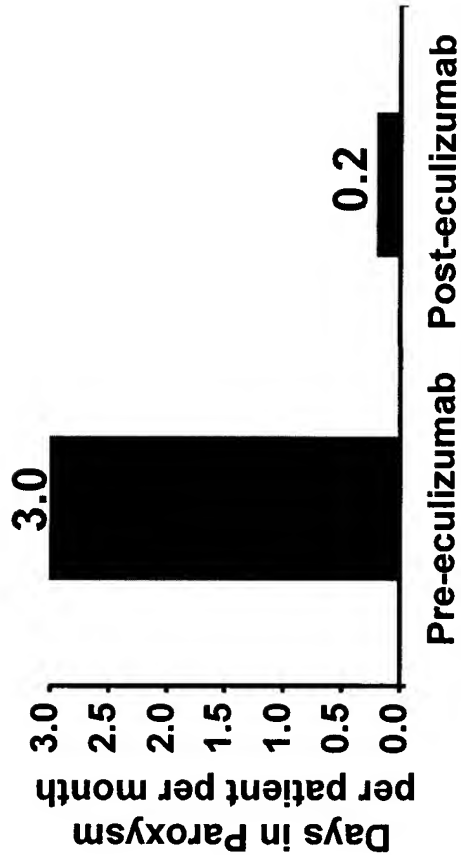


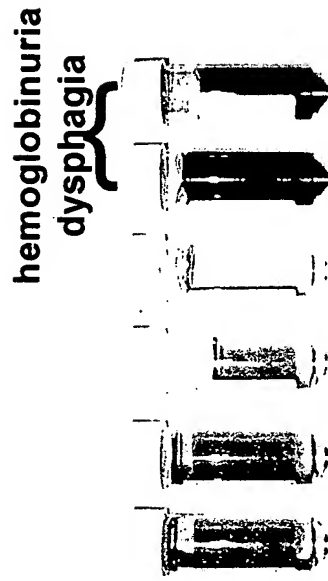
Figure 2

Figure 3: Effect of Eculizumab on Paroxysm Rate (n=8)



As compared to the number of paroxysms observed in patients during the screening period prior to eculizumab therapy, the number of events was reduced 93% from 3.0 days of paroxysms/patient/month to 0.2 days of paroxysms/patient/month ($p<0.001$) during the one year of eculizumab therapy

Fig 4. Early Morning Urine



Visit #	4								5	9
Days	0*	1-8	9	10	11	12	13		0*	1
Urine	10	2-3	3	3	3	3	9		10	3
LDH	2624	-	784	-	-	697	1687		2917	-
AST	119	-	38	-	-	31	87		-	-
PD	61	-	16	-	-	5	30		45	-

PD, % serum hemolytic activity; values under 20% are considered completely blocked; *Dose of eculizumab

Fig 6b: Break-through of complement blockade resulted in hemoglobinuria, dysphagia, and increased LDH and AST. At the next dose, symptoms resolved and reduction from 900mg every 14 days to 900mg every 12 days resulted in a regain of complement control which has been maintained for over 9 months in both patients (See Fig 6c). Both episodes were well tolerated without evidence of serious adverse events.

Fig 5. Patient 010-010 Urine Grade vs. Time

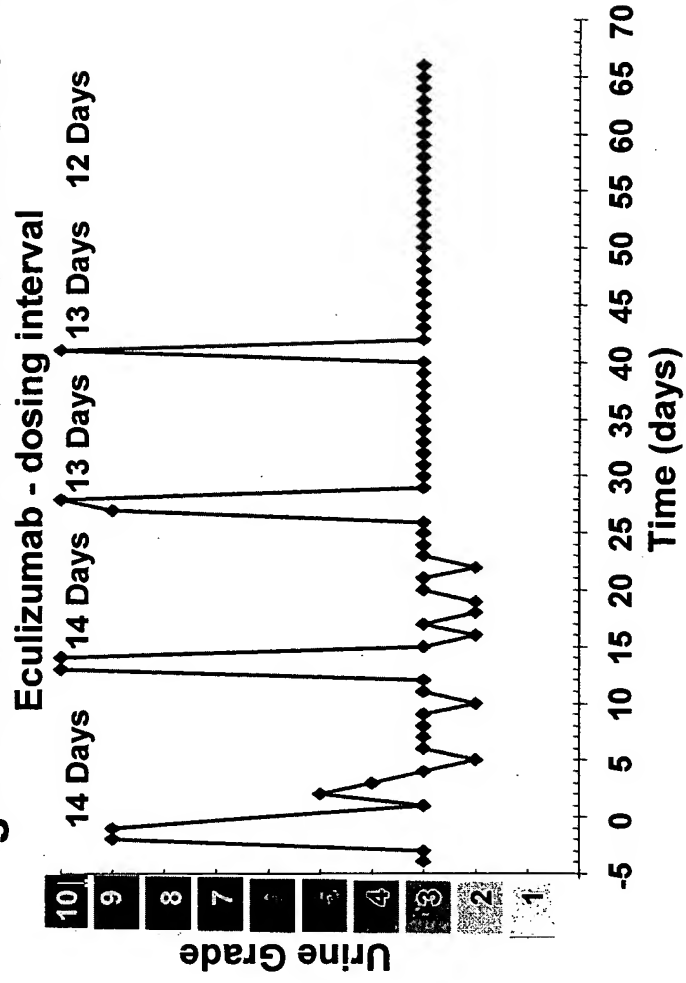
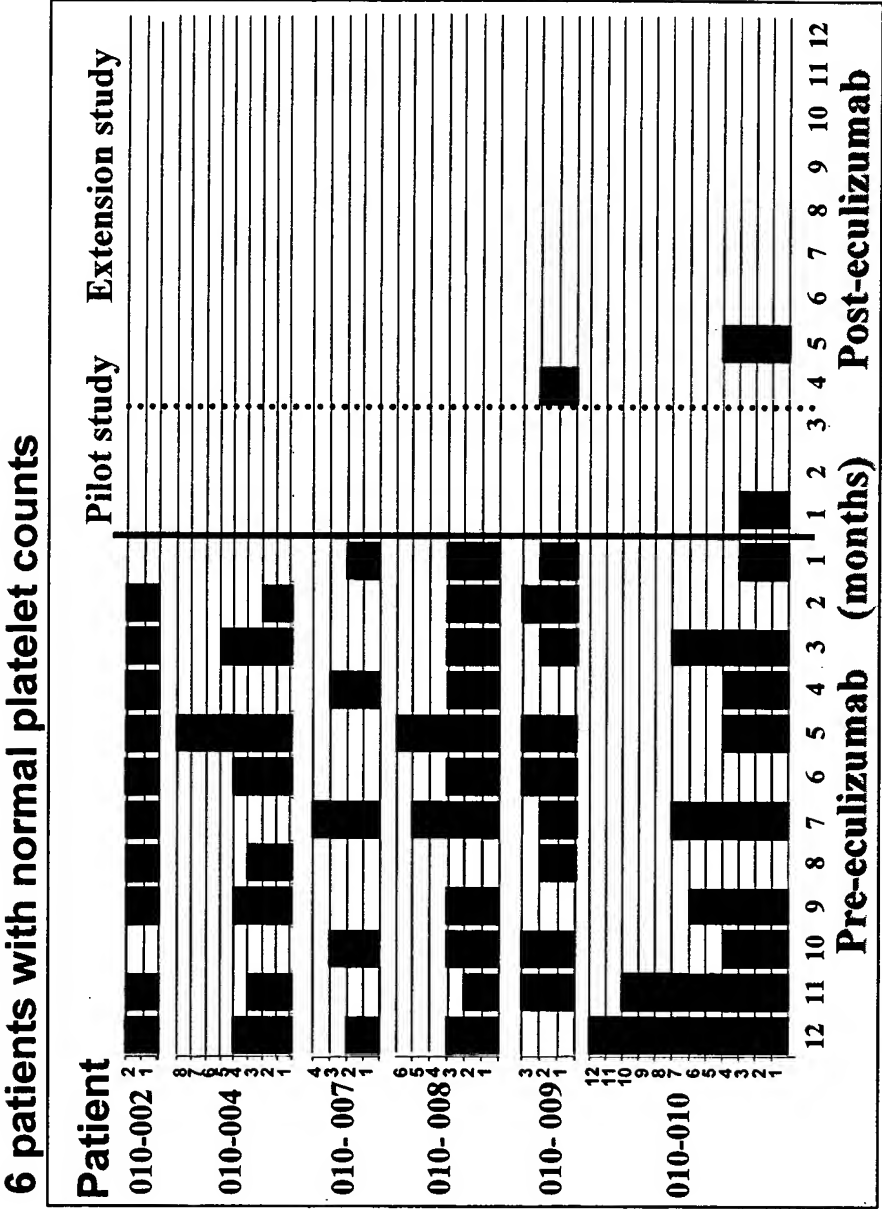
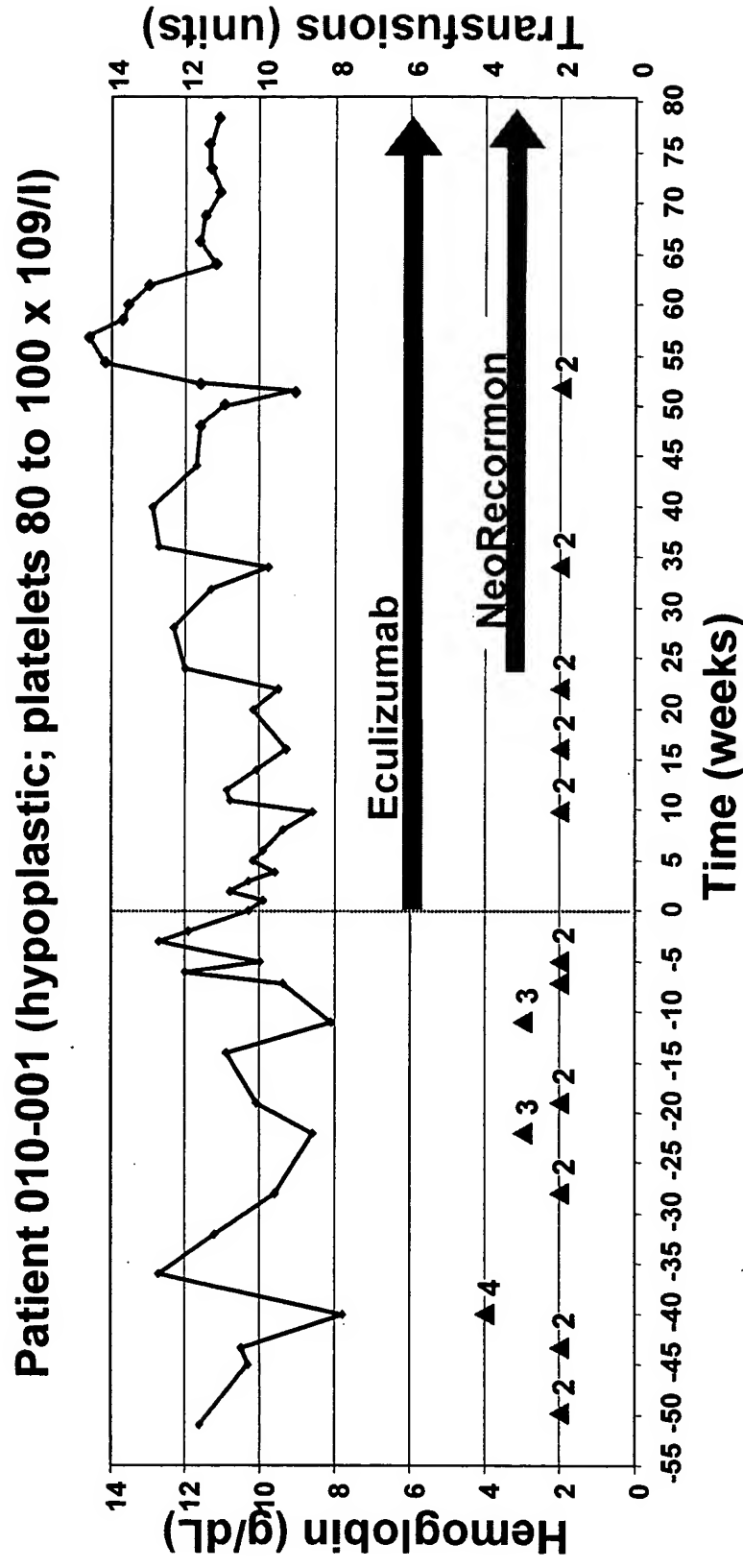


Figure 6: Transfusions

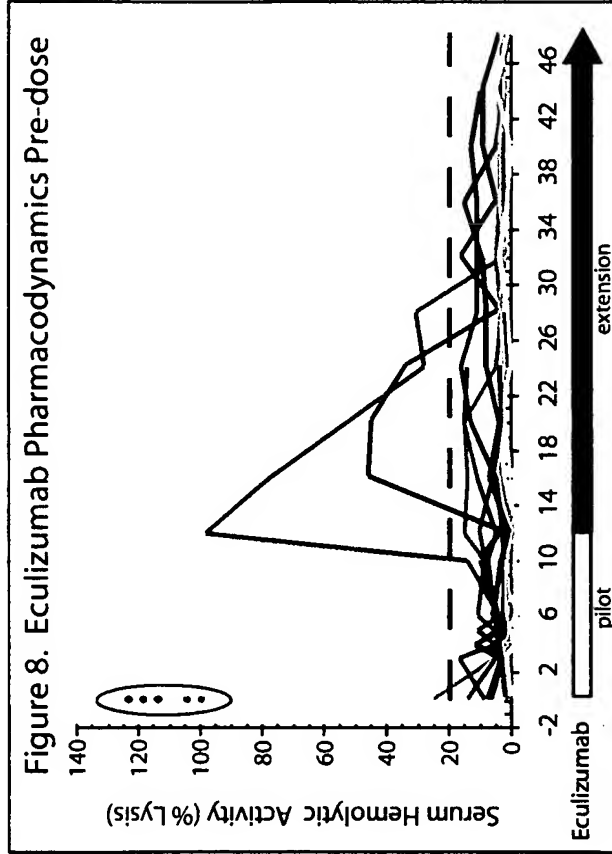


Non-thrombocytopenic patients have better response and most become transfusion-independent

Figure 7 Management of thrombocytopenic patient with erythropoietin



Transfusions reduced with eculizumab and transfusion independence with combined eculizumab and erythropoietin (NeoRecormon 18,000U 3x week)



Nine out of eleven patients were completely blocked for longer than 12 months and 2 patients (010-010 & 011-001) transiently escaped from complement blockade just prior to doses of eculizumab .

Figure 9: Quality of Life During One Year of Eculizumab

QOL Parameter (a)	Baseline (b)	Change from Baseline (c)	p-value
Global Health Status	56.1	14.5	0.005
Physical Functioning	70.9	13.9	<0.001
Role Functioning	66.7	14.2	0.005
Emotional Functioning	70.5	13.1	<0.001
Cognitive Functioning	77.3	11.4	<0.001
Fatigue	47.5	-17.7	<0.001
Pain	21.2	-7.6	0.032
Dyspnoea	39.4	-15.4	<0.001
Insomnia	30.3	-8.7	0.023

a) Quality of life assessed using the EORTC QLQ-C30 instrument; b) Numbers represent mean values of transformed scores; c) Mixed Effect Ancova Model: Positive change indicates improvement for Global Health Status, Physical Functioning, Role Functioning, Emotional Functioning and Cognitive Functioning while a negative change indicates improvement for Fatigue, Pain, Dyspnoea and Insomnia